

CONGENITAL MUSCULAR DYSTROPHIES

The congenital muscular dystrophies (CMDs) are a group of inherited muscle disorders that typically become apparent at birth or in early infancy and are characterised by floppiness, weak muscles or problems with the mobility of joints.

Symptoms in small babies include hypotonia (floppiness) and low muscle tone. As the muscles are weak and immobile, contractures (tightness) in the hip, ankle, knee and elbow joints are common. In children that do not have contractures, initial problems may be difficulties holding the head, and delays in sitting and walking.

Duchenne muscular dystrophy (DMD) is a muscle wasting condition. Muscle weakness occurs because of a genetic defect that prevents dystrophin (a protein in muscle) functioning correctly. The affected gene resides on the X chromosome, so only boys are affected. The first signs of muscle weakness typically occur before the age of four years, for example difficulties rising from the floor, walking, running, jumping and climbing stairs. Wheelchair users are at risk of developing spinal curvature, which can develop into scoliosis. DMD boys may have learning or behavioural difficulties, their heart or respiratory muscles may be affected.

Becker muscular dystrophy (BMD) is a rare genetic condition associated with mutations (changes) in the dystrophin gene. A reduction in the quantity or functioning of dystrophin leads to damage to the muscles. BMD largely affects males.

Spinal muscular atrophy (SMA) is a rare, inherited muscle-wasting condition, which may affect a child's ability to crawl and walk, to move arms, hands, head and neck, and can affect breathing and swallowing.

*Children's needs are **individual** - some children may require additional support:*

- Fine motor skills, e.g. hand skills including two-handed tasks such as cutting, grasping and releasing objects
- Gross motor skills possibly requiring a differentiated PE curriculum
- Maintaining body posture, sitting may need specialist seating
- Mobility, e.g. moving around the classroom, learning environments
- Self-help skills, aspects of personal care, e.g. toileting, dressing, feeding
- Tiring during the day, due to increased effort
- Emotional support to enhance self esteem, particularly in peer groups, coming to terms with the implications of the condition, 'losing skills' previously attained

FURTHER INFO: Muscular Dystrophy Campaign www.muscular-dystrophy.org